Traitement chirurgical des arthropathies hémophiliques

C. Bresson [a,*], M. Dauty [b]

[a] *Médecine légale, CHU de Nantes, place Alexis-Ricordeau, 44093 Nantes cedex 1, France*

[b] *CHU de Nantes, Nantes, France*

*Auteur correspondant.

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La répétition des hémarthroses provoque des lésions articulaires responsables de retentissements fonctionnels variables d'un patient à l'autre. Comme pour les pathologies dégénératives habituelles, il y a peu de parallélisme anatomo-clinique. En conséquence, l'indication chirurgicale ne dépend pas uniquement de l'évolution radiographique, mais essentiellement de la plainte fonctionnelle. L'importance de la dégradation articulaire constatée sur les arthropathies hémophiliques, (notamment, le coude, le genou ou la cheville) conduit le chirurgien a réalisé souvent des arthrodèses ou des arthroplasties prothétiques.

L'acte chirurgical impose bien évidemment une prise en charge multidisciplinaire. L'importance des raideurs existantes et surtout leur chronicité, nécessite d'associer un travail de rééducation souvent en préopératoire et systématique en postopératoire. Le coût du traitement substitutif en facteurs de coagulation impose une localisation géographique dans un centre référent.

Les prothèses sont essentiellement utilisées sur la hanche, le genou et le coude ; les arthrodèses sont souvent préférées sur l'articulation de la cheville.

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Version anglaise

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Why do joints bleed in hemophilia?

M. Trossaert

Centre régional de traitement de l'hémophilie, 30, boulevard Jean-Monnet, 44093 Nantes, France

Keywords: Hemophilia; Hemophilic arthropathy

The coagulation process can be activated by two pathways: the cascade reactions can be initiated by the contact phase (intrinsic pathway) and/or the presence of tissue factor (TF) can induce coagulation via the extrinsic pathway. The injured tissue induces release of TF, a phenomenon now confirmed as the main process activating coagulation, leading to maximal thrombin generation and fibrin clot formation. However, the two anti-hemophilic factors (Factors VIII and IX) do not belong to this extrinsic coagulation pathway. Thus, it was initially difficult to explain why patients with hemophilia (in whom the main coagulation pathway is preserved) do suffer from hemorrhagic symptoms.

In the 1970s, François Josso et al. demonstrated that the first steps of the coagulation activation always require factor V and Factor VIII activation (now defined as the "Josso loop"), that thrombin generation plays a major role in this process and suggested that another coagulation pathway could be critical to explain the predisposition of joints to bleed in patients with hemophilia. Since this first description, it has been demonstrated that TF can be identified in many tissues (epidermal tissues, digestive mucosa, aero-digestive tract), but not in plasma, normal blood cells, in joint tissue and striated muscle. So, when there is no TF, the extrinsic pathway becomes the major pathway and its dysfunction could easily lead to abnormal bleedings. These locations are then predisposed to bleed in hemophilic patients.

In the absence of prophylaxis, the hemophilic arthropathy often takes place very early in children with severe hemophilia, mainly located in knees, ankles and elbows. Hemarthrosis is usually accompanied by rapid pain, joint swelling and muscular dysfunction. With adapted substitution treatment (Factor VIII or IX concentrates), these clinical signs disappear in a few days. In contrast, the repetition of these joint bleedings induces a progressive destruction of the cartilage and certain synovia alterations. This joint destruction is the first cause of morbidity that can alter the quality of life of hemophilia patients.

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The clinical course of the haemophiac joint

M. Dauty

MPR locomoteur, CHU de Nantes, hôpital Saint-Jacques, 85, rue Saint-Jacques, 44035 Nantes cedex 01, France

Keywords: Haemophilia; Joint; Rehabilitation

Objective.– Haemophilia is responsible for joint bleedings because of coagulation factor deficit. When bleedings are repeated, a vicious circle ends in a progressive and complete destruction of joints. The purpose of this lecture is to show how this destruction occurs clinically while focusing on various preventive therapeutic options.

Method.– After a brief reminder of the physiopathological factors of haemophiliac joint diseases, various joint topographies are described. Clinical and radiological evolution is shown from clinical cases. Various clinical scores are also discussed. The clinical course of the joint is discussed, according to preventive treatment with anti-haemophilic factors and according to the presence of an inhibitor. The various conservative treatments used at different stages of joint destruction are exposed.

Results.– Three joints, elbows, knees and ankles have a particularly high risk of destruction. According to the possibilities of disease prevention, 1 to 3 joints will be destroyed by the age of 20 years versus 5 if no preventive treatment is given. The presence of an inhibitor is a powerful protection for joint integrity. Physical medicine and rehabilitation treatments are used to restore deficiencies and to improve pain relief. Synoviorthesis attempts to limit the synovial proliferation which evolves on the own and participates in the joint degradation.

Discussion.– Preservation of joints in haemophiliac subjects depends on disease prevention with anti-haemophilic factors but also on exposure to risk resulting from proposed activities and compliance with an annual joint follow-up to guarantee early treatment of haemarthrosis and early-stage arthropathy. This specific assessment requires multidisciplinary medical cooperation to protect joint integrity throughout the life of the haemophiliac subject.

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Surgical treatment of hemophilic arthropathy

C. Bresson [a], M. Dauty [b]

[a] *Médecine légale, CHU de Nantes, place Alexis-Ricordeau, 44093 Nantes cedex 1, France*

[b] *CHU de Nantes, Nantes, France*

Keywords: Hemophilic arthropathy; Surgery

Repeated hemarthrosis causes joint damage responsible for functional impairment variable from one patient to another. As usual with degenerative diseases, there are no predicting factors on radiographs. Consequently, the indication for surgery depends not only on radiographic progression, but essentially the functional complaint.

The importance of joint damage seen in hemophilic arthropathy (especially elbow, knee or ankle) often leads the surgeon to perform arthrodesis or prosthetic replacements. The surgery obviously requires a multidisciplinary approach. The significant stiffness and especially its chronicity, requires systematic post-operative rehabilitation and sometimes preoperative rehabilitation. The cost of clotting factors implies a geographic location near a referral center. Prostheses are mainly used for the hip, knee, and elbow; arthrodesis is often preferred for the ankle joint.

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